

# Juvenile Cystic Adenomyoma Mimicking a Uterine Anomaly: a Report of Two Cases

*Bir Uterin Anomaliyi Taklit Eden Jüvenil Kistik Adenomyom: İki Vakanın Sunumu*

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## ABSTRACT

Juvenile cystic adenomyoma is a rare form of adenomyosis and described as a new type of müllerian anomaly in literature. We are presenting two cases of juvenile cystic adenomyoma which were misdiagnosed preoperatively as unicornuate uterus with haematometra in a non-communicating rudimentary horn. The mainstay of treatment is complete resection of lesion.

**Keywords:** Juvenile cystic adenomyoma, müllerian anomaly, unicornuate uterus

## Öz

Jüvenil kistik adenomyom nadir bir adenomyoz şeklidir ve literatürde yeni bir müllerian anomali türü olarak tanımlanmaktadır. Bu çalışmada ameliyat öncesinde nonkomünike rüdimenter horn'da hematometralı unikornuat uterus olarak yanlış tanı konulan jüvenil kistik adenomyomlu iki vaka sunulmaktadır. Tedavinin temeli lezyonun tamamının rezeksiyonudur.

**Anahtar Kelimeler:** Jüvenil kistik adenomyom, müllerian anomali, unikornuat uterus

## Introduction

Juvenile cystic adenomyoma (JCA) is a rare form of adenomyosis. It affects young girls and usually manifests as severe dysmenorrhea and recurrent pelvic pain. Patients are often misdiagnosed to have müllerian anomalies such as a unicornuate uterus with hematometra in a non-communicating rudimentary horn or a bicornuate uterus with segmental atresia because of the typical clinical presentation and the rarity of the condition. In this paper, two cases of JCA are presented.

## Case Reports

### Case I

A 23-year-old unmarried female presented with on-and-off severe episodic pain in the lower abdomen for 2 months and severe dysmenorrhea for 3 years. The pain was spasmodic in nature, non-radiating, not associated with any urinary and gastrointestinal symptom, and was refractory to analgesics. She attained menarche at 17 years of age. Her menstrual cycles were regular with average flow. Minimal tenderness was present over the lower abdomen on the right side. Trans-abdominal ultrasound (TAS) showed a normal-sized uterus with hematometra (3.9×2.9×3.9 cm) in the anterior wall of the uterus near the cornual end, which was suggestive of a unicornuate uterus with a non-communicating rudimentary horn (Figure 1). Informed and written consent was obtained for performing surgery. On performing diagnostic laparoscopy, the uterus was 6-8 weeks in size with a bulge seen in the right anterior wall near the cornual end; the right tube was not attached on this bulge. On performing hysteroscopy, the uterine cavity was normal, and bilateral ostia were seen. On laparoscopic guided needle aspiration of the bulge, chocolate-colored blood was aspirated, suggestive of an adenomyotic cyst. A decision for laparotomy was taken and the abdomen was entered through a Pfannenstiel incision. An incision was made on uterus over the bulge, thick chocolate-colored fluid was drained, whole cyst wall (4×4cm) was excised and dead space obliterated with 1-0 vicryl suture. A histopathological examination confirmed the diagnosis of an adenomyotic cyst. Her pelvic pain and dysmenorrhea were completely resolved after surgery. She was prescribed combined oral contraceptives pills (OCPs) for 1 year;



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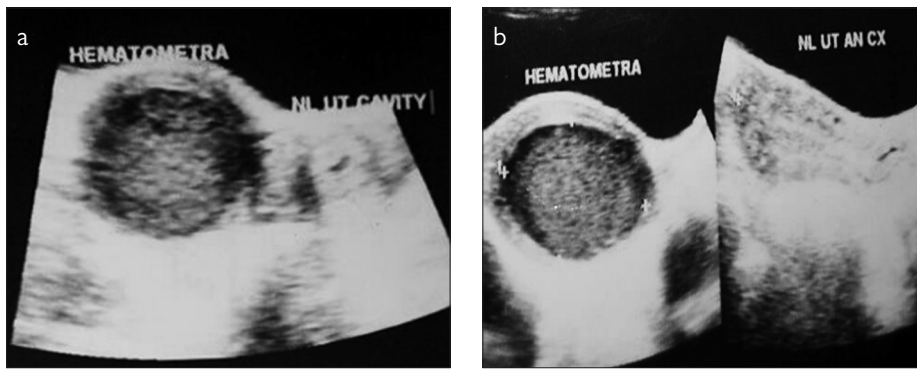
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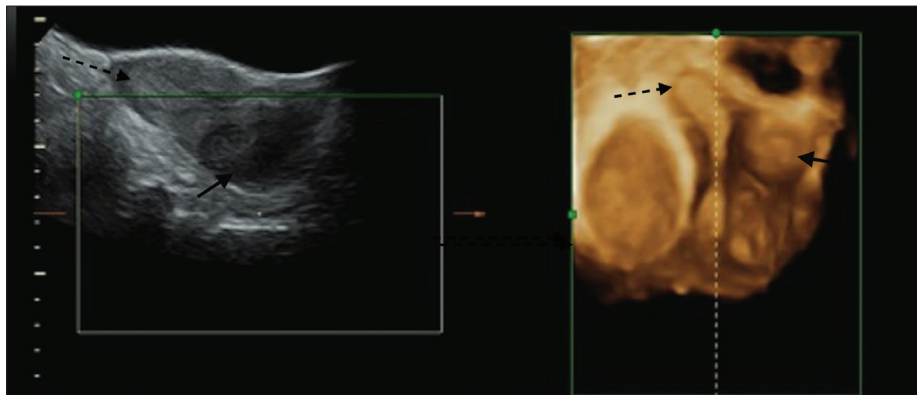
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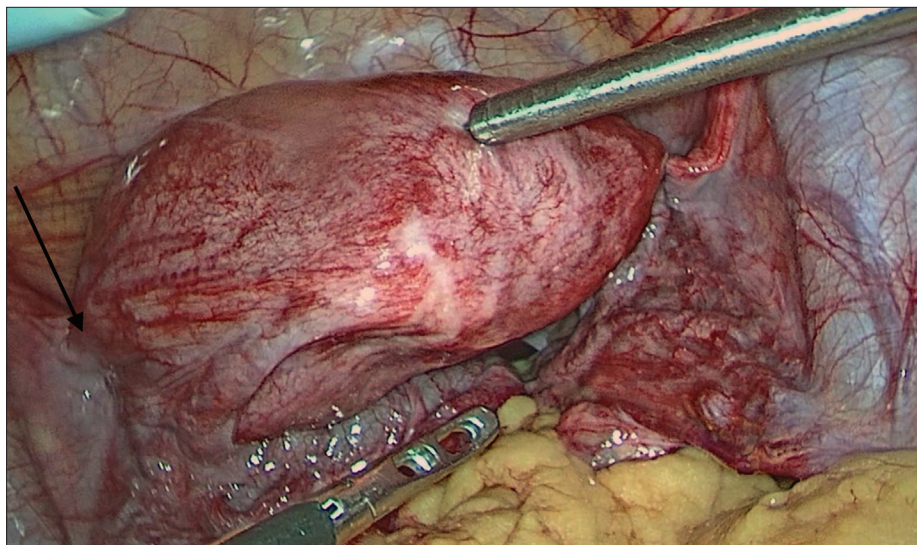
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**Figure 1. a, b.** TAS shows a well-circumscribed lesion in the right uterine myometrium (hematometra); a normal uterine cavity is separately seen in the (a) transverse view and (b) longitudinal view.



**Figure 3.** 2D and 3D USG images show a well-circumscribed lesion in the anterior myometrium (solid arrow); the uterine cavity (broken arrow) is seen separately.



**Figure 4.** Intraoperatively, a bulge is seen over the left side of the anterior uterine myometrium near the insertion of the round ligament (Arrow).

to prevent recurrence. She conceived 3 years after surgery and gave birth to a healthy baby.

## Case 2

A 16-year-old unmarried girl presented to us with acute episodic pain in the left lower abdomen; the pain was severe in nature and occurred every 2-3 months for the last 3 years since men-

arche. Her menstrual cycles were regular with an average flow; she had severe dysmenorrhea. She required analgesics for pain relief. She had already undergone ultrasound and magnetic resonance imaging (MRI). Both TAS and MRI were suggestive of a unicornuate uterus with hematometra in the left non-communicating rudimentary horn (Figure 2). On reviewing the



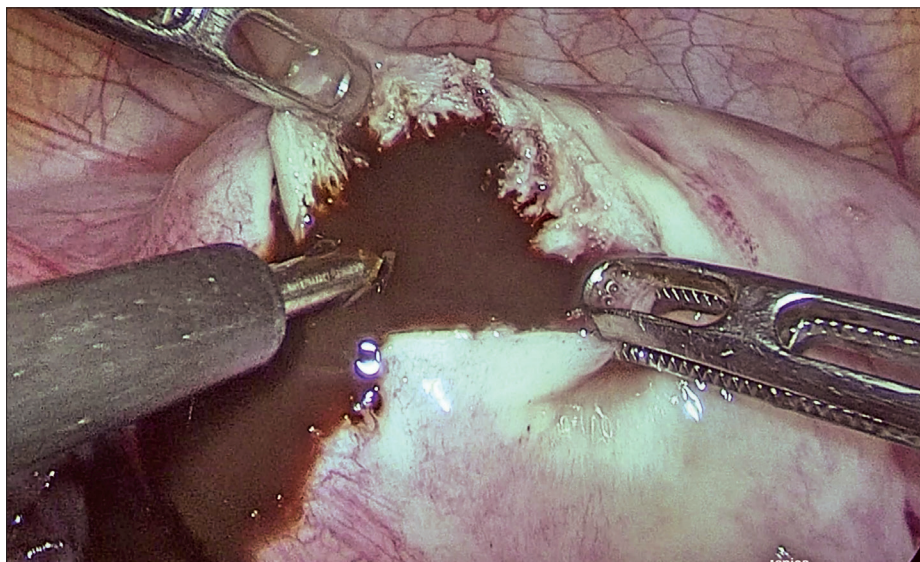
**Figure 2.** T1-weighted MRI film in the sagittal section demonstrates a well-circumscribed lesion in the anterior myometrium (solid arrow), which is separately seen from the uterine cavity (broken arrow).

MRI films and performing a three-dimensional scan, JCA was suspected. Informed and written consent was obtained (Figure 3). On performing laparoscopy, the uterus appeared unevenly enlarged, with a bulge that was 4×3cm over the left uterine wall near the cornual end just below the insertion of the round ligament; there was no rudimentary horn (Figure 4). The bilateral tubes and ovaries appeared healthy. Diagnostic hysteroscopy was suggestive of a normal-sized uterine cavity and well-visualized bilateral ostia. A transverse incision was made over the bulge, and thick chocolate-colored fluid was drained (Figure 5). The entire adenomyotic cyst with surrounding adenomyotic tissue was removed and sent for a histopathological examination, which confirmed the diagnosis of an adenomyotic cyst (Figure 6). The uterine incision was closed with I-0 V-lock suture. The patient was advised to take low-dose OCPs for hormonal suppression. Postoperatively, she is doing well.

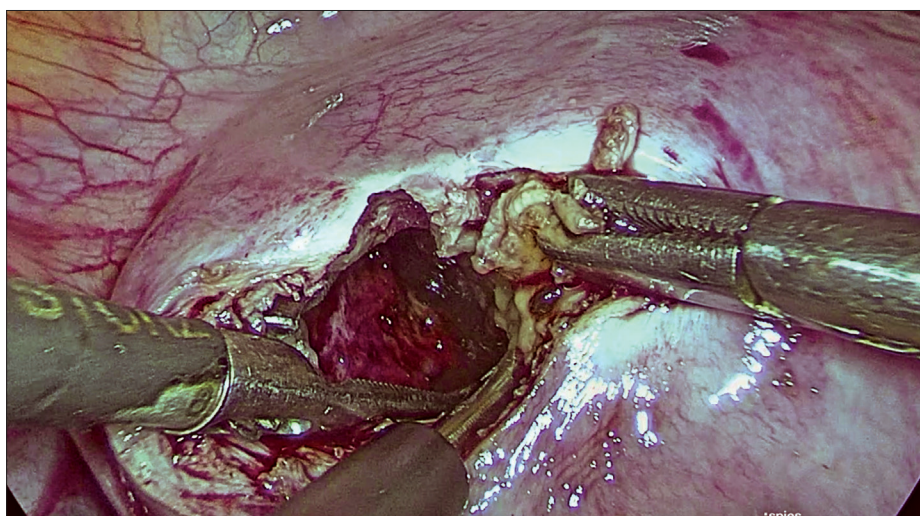
## Discussion

The actual incidence of JCA is unknown. Because of improved imaging techniques and increased awareness, many cases of JCA are now being reported. These lesions are known by several names such as JCA, juvenile adenomyotic cyst, and recently, as an accessory and cavitated uterine mass [1-3]. Its exact etiology is unknown. Acien et al. [1] proposed that these lesions should be considered as a new type of congenital müllerian anomaly because of early onset dysmenorrhea and their specific location at the anterior uterine wall near the attachment of the round ligament with the uterus. These lesions should be separately classified, as





**Figure 5.** Chocolate-colored fluid drained from the lesion.



**Figure 6.** Adenomyotic cyst cavity.

unlike other müllerian anomalies, the uterine cavity is normal in JCA. These lesions can be caused by the duplication of ductal müllerian tissue at the level of attachment of the round ligament, which is possibly related to gubernaculum dysfunction [4]. These findings were present in both our cases. Takeuchi et al. [5] considers the juvenile form as a cystic variant of adenomyosis rather than as a congenital anomaly.

The most appropriate diagnostic criteria were provided by Takeuchi et al. [5]; these included age of 30 years or younger; cystic lesion  $\geq 1$  cm in diameter and independent of the uterine lumen and covered by hypertrophic myometrium on imaging or intraoperatively and associated with severe dysmenorrhea.

Though the MRI results were misinterpreted by our radiologist, MRI is the imaging modality of

choice as it is non-invasive and clearly shows the pelvic anatomy, cavitated mass with hemorrhagic contents, and the uterus, myometrium, and endomyometrial interface [2].

In our first case, we suspected an adenomyotic cyst as the contour of the uterus did not suggest a rudimentary horn on performing laparoscopy; hysteroscopy was useful in confirming the diagnosis. In the second case also, although MRI reported a rudimentary horn we suspected JCA on 3D ultrasound (USG) which was confirmed on hysteroscopy. In case of doubt, hysteroscopic visualization of both uterine ostia can rule out an obstructive müllerian anomaly.

Medical management options include non-steroidal anti-inflammatory drugs, continuous or cyclical OCPs, and gonadotropin-releasing hormone analogs. However, the role of medical

management is limited and provides only temporary and partial relief [5].

The gold standard treatment for JCA is complete resection of the lesion. Laparoscopy has been performed as a minimally invasive method [5, 6].

To conclude, many cases reported in the literature were preoperatively misdiagnosed as müllerian anomalies, cystic degeneration in adenomyomas and leiomyomas, and broad ligament fibroids. Gynecologists, as well as radiologists, should be aware of and keep a high index of suspicion about this condition to make an accurate pre-operative diagnosis. JCA is a treatable cause of severe dysmenorrhea. It should be kept in mind as a differential diagnosis in young girls with severe dysmenorrhea.

**Informed Consent:** Verbal informed consent was obtained from patient who participated in this study.

**Peer-review:** Externally peer-reviewed.

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